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Dermatofibrosarcoma Protuberans Metastasizing to Lymph Nodes: A Case Report and Review of Literature

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Dermatofibrosarcoma Protuberans (DFSP) is an uncommon cutaneous soft tissue tumor. It is locally invasive and is known to recur. Metastases are rare and occur most commonly to the lungs. Metastasis to lymph nodes is extremely rare with only a few case reports in literature. The management strategy for such lymph node metastases remains controversial because of rarity of the condition. We report a case of DFSP of the lower limb with definite inguinal node secondaries managed by loco-regional surgery. We recommend that block dissection of lymph nodes should be performed in all cases with secondaries.
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KEY WORDS: dermatofibrosarcoma protuberans; fibrohistiocytic tumor; sarcoma

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is an uncommon cutaneous soft-tissue tumor. It is locally invasive and is known to recur. Metastases are rare and occur most commonly to the lungs. Metastasis to lymph nodes is extremely rare, with only a few case reports in literature. The management strategy for such lymph node metastases remains controversial because of rarity of the condition. We report a case of DFSP of the lower limb with definite inguinal node secondaries managed by locoregional surgery.

CASE REPORT

A 30-year-old otherwise healthy man had recurrent swelling in the back of his left thigh for the 2 years previous to presentation. The patient had had a similar swelling 4 years earlier, which had been excised completely at another medical center; no record of histopathologic diagnosis was available. Examination revealed a 5 × 5 cm hard cutaneous swelling in the mid-thigh posteriorly (Fig. 1A). The swelling was mobile over underlying muscles, with evidence of previous scar and areas of altered pigmentation. No other satellite skin or subcutaneous nodes were present. There was a large ipsilateral inguinal lymph node (6 × 5 cm) with four smaller discretely palpable nodes (Fig. 1B). They were firm in consistency and there was no fixation to overlying skin or underlying tissues. Abdominal examination was normal. The clinical diagnosis was melanoma or a soft tissue tumor. Fine-needle aspiration cytology of the primary tumor and the inguinal lymph nodes showed features suggestive of DFSP at both sites.

The patient had radical surgery; the primary tumor was excised with a margin of 5 cm, including the underlying fascia and muscles, and inguinal lymph nodes were block-dissected. The primary defect was covered by a split-thickness skin graft. The patient made an uneventful postoperative recovery except for a small area of marginal flap necrosis (2 × 1 cm) in the left inguinal region, which healed conservatively. The histopathologic examination confirmed the diagnosis of DFSP at the primary site with metastases in the lymph nodes (Fig. 2). The patient is asymptomatic 2 years after surgery.

DISCUSSION

DFSP is classified as a fibrohistiocytic tumor of intermediate malignancy [1]. The tumor infiltrates locally and

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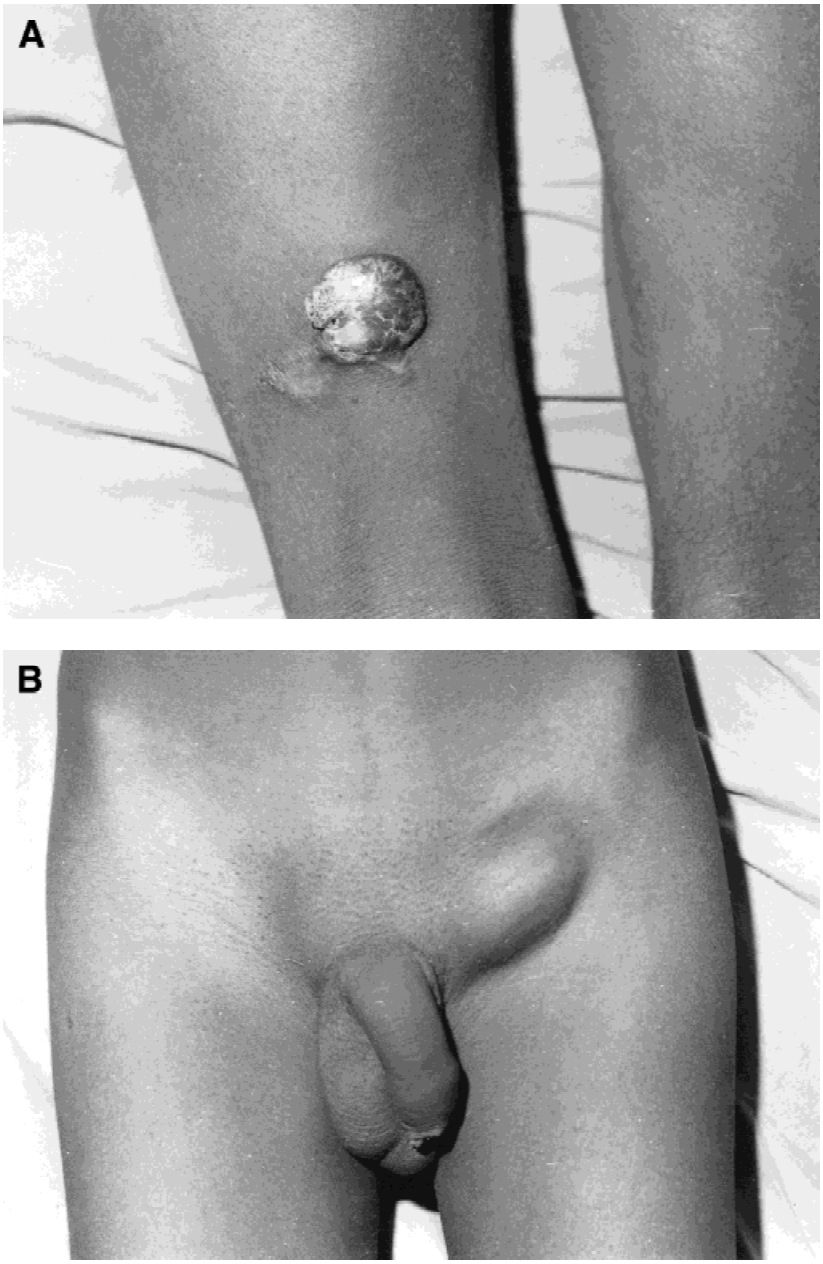

Fig. 1. **A:** Primary tumour on the posterior aspect of the left thigh. Note the areas of altered pigmentation and scar of the previous surgery. **B:** Enlargement of left inguinal lymph nodes.

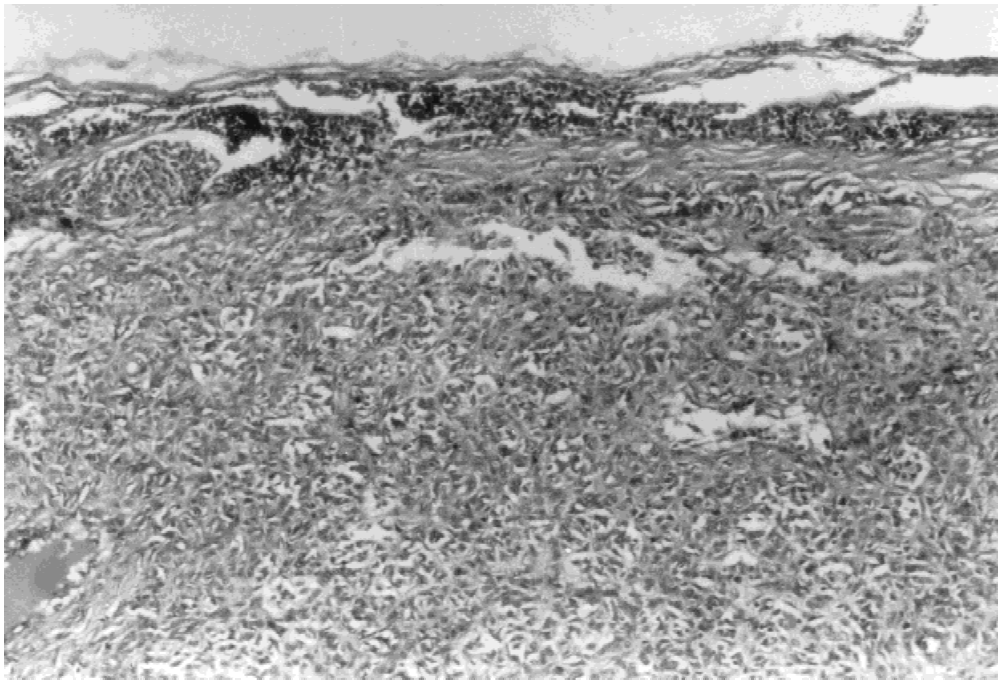

Fig. 2. Photomicrograph of a lymph node showing a uniform population of fibroblasts arranged in a monotonous storiform pattern lying in the subcapsular sinus. Note the capsule of the lymph node and the lymphoid cells pushed to the periphery. Hematoxylin-eosin, ×100.

only very rarely metastasizes. It recurs in up to half of the patients within 3 years of the initial surgery [2]. Metastases, though uncommon, are predominantly to the lungs. Metastasis to lymph nodes is largely unknown, and only a few controversial case reports are available in the world literature. To date, only 16 cases of metastatic disease have been reported, of which only 4 had documented lymph node metastasis [3]. One out of these 4 cases was unique in that the lymph node metastases were likened to Hodgkin's lymphoma [4].

This lesion has to be differentiated mainly from the nodular variety of malignant melanoma, which resembles metastatic DFSP in clinical appearance with regard to palpable lymph nodes. Accurate diagnosis can be made by fine-needle aspiration cytology of the primary tumor and the secondary lesions in the lymph nodes. The mode of treatment has not been well described in literature because of the extreme rarity of this condition. Wide local excision with block dissection of lymph nodes is recommended as the primary mode of management of DFSP with lymph node metastases.

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